BENIGN AND MALIGNANT CHORDOMAS
A CLINICO-ANATOMICAL STUDY OF TWENTY-TWO CASES*

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It is convenient to divide chordomas into benign and malignant types. The benign lesions have been found only at necropsy except for the benign coccygeal chordoma included in this report, which was found by chance in a surgical specimen. The relative inaccessibility of the asymptomatic benign tumor accounts for this scarcity of case reports in comparison to the several hundred malignant chordomas that have been described in the literature.[1,2]

BENIGN CHORDOMAS

A summary of four benign chordoma cases follows:

Case 1

A 68-year-old man was admitted to the University Hospital with hypertensive arteriosclerotic heart disease and prostatism. He died suddenly 5 days following a minor surgical operation. No symptoms or physical findings relative to the base of the brain, except for bilateral nerve deafness, were elicited. At necropsy the findings of arteriosclerotic heart disease were demonstrated. In addition, a section through the pituitary gland showed a mass of physaliferous cells outside the circular sinus in the sulcus between the anterior and posterior lobes (Fig. 1). Serial sections showed the chordoma cells outside the circular sinus throughout the thickness of the paraffin blocks.

Case 2

A 32-year-old woman died 24 hours after an osteoplastic craniotomy for a spongioblastoma polare of the right side of the brain stem and the right thalamus. A necropsy limited to the head showed the remainder of the glioma and also a benign chordoma which was loosely attached to the ventral surface of the pons. No attachment to the clivus was noted. Microscopically the chordoma was composed of large vacuolated cells with little intercellular mucin.

Case 3‡

A 61-year-old woman in an institution because of involutional psychosis died of coronary thrombosis. At necropsy the brain stem showed

a gray-white, gelatinous nodule, 0.8 cm. in diameter, which had a very loose attachment to the right anterior surface of the pons. No dural attachment was noted. Vacuolated and non-vacuolated cells made up the neoplasm and cords of cells were separated by intercellular mucin.

Case 4

A 44-year-old man had rectal symptoms referable to hemorrhoids. Through the sigmoidoscope a specimen for biopsy was obtained from a lesion on the anterior rectal wall, 11 cm. above the anus. This proved to be a carcinoid. Coccygectomy was done to facilitate removal of a segment of rectum. A routine section of the coccyx showed a benign chordoma arising in the first intercoccygeal disk and growing beneath the periosteum on the posterior surface of the coccyx (Figs. 2, 3, and 4). Serial sections of the coccyx showed the tumor growing along the surface of the disk on the right side of the coccyx and nearly surrounding it. It did not extend completely through the periosteum.

DISCUSSION

Virchow[3] and Luschka[4] were first to describe the benign chordoma on Blumenbach's clivus. They interpreted it as a chondromatous neoplasm, and ecchondrosis physalifora spheno-occipitalis was used by Virchow and his followers to designate the lesion. Müller[5] and Ribbert[6] established the origin of this neoplasm from notochordal tissue and the name was changed later to ecchordosis physalifora spheno-occipitalis. Stewart and Burrow[1] reported 4 cases arising from Blumenbach's clivus.

With the demonstration of collections of histologically similar cells having the features of benign neoplasms at different levels along the original tract of the notochord from the pituitary region to the coccyx, it seems unnecessary to continue to use the cumbersome term ecchordosis physalifora and better to refer to the lesion as a benign chordoma, particularly when it occurs in adults. The term notochordal rest has some usefulness in designating collections of physaliferous cells in fetuses and young children which persist in the vertebral portion of the notochordal canal and which lie along the irregular branches of the original notochord. The latter can be referred to also as ectopic notochordal rests or chordal ectopia after Horwitz.[2]

The physaliferous cells of the notochord which give rise to the nucleus pulposus and which can be found in that structure until about the seventh year of life are referred to in this paper as nucleus pulposus cells.[7] Some workers have described these cells in later life in the

intervertebral disk and sacrum.[8,9] The acquired Schmorl's nodule and the posteriorly herniated nucleus pulposus have no special relation to the chordoma problem since these retrogressive lesions do not contain nucleus pulposus cells except in rare instances.[8] Eckert and Decker[10] failed to find nucleus pulposus cells in adult cadavers or in herniated intervertebral disks.

In spite of the limited number of case reports, a fairly high incidence of benign clival chordomas has been determined. Ribbert[11] found 10 cases in 500 necropsies or an incidence of 2 per cent. Stewart and Burrow[1] found 3 examples in 200 necropsies or a 1.5 per cent incidence. The incidence has not been determined for the pituitary region, the dens epistropheus, or the coccyx. From Schmorl's collection of spines Beadle[12] reported 7 cases of persistent notochordal cells which can be interpreted as benign chordomas. Six were in dorsal vertebrae and one was in the first lumbar vertebra.

The benign chordoma is an asymptomatic lesion. On Blumenbach's clivus the small, soft tumor is circumscribed and has not been shown to give pontine or bulbar symptoms. A thin pedicle attached through a defect in the dura to the underlying bone is found in most cases but was not observed in the cases reported here. The cells making up the neoplasm are quite uniform. They may or may not have vacuolated cytoplasm and the amount of intercellular mucin varies, but large pools of mucin often found in the malignant chordomas were not found in these 4 cases. Neoplasm size, invasiveness, mitotic figures, pleomorphism, excessive mucin formation, and other features of malignancy readily separated these benign neoplasms from the malignant cases. The similar location and cell type of the notochordal rests, nucleus pulposus cells, the benign chordoma, and the malignant chordoma indicate a close relationship of these four cellular configurations. Ribbert[13] and Congdon[14] have studied this problem experimentally and produced a lesion in rabbits by puncturing the nucleus pulposus which morphologically resembled a chordoma. True malignancy was not observed.

MALIGNANT CHORDOMAS

The malignant chordoma is considered a rare neoplasm and for this reason is a diagnostic problem. A general survey, however, of all primary supporting tissue neoplasms of the vertebral column seen in the Department of Pathology indicated that it formed a significant percentage of this group, particularly if neoplasms of the spinal cord and its meninges were excluded. A brief summary of each case follows.

Case 5

A 13-year-old boy was struck on the head by a stone 2 years before admission. Following this he had a stiff neck, with pains in head and neck. Five weeks before admission he developed nausea, vomiting, dizziness, and convulsions. Physical examination showed involvement of the left 11th and 12th cranial nerves. There was a scar on the occiput from the original injury. Roentgenologic examination showed a normal skull and cervical spine. A clinical diagnosis of posterior fossa tumor in the left cerebellar region was made. Death occurred during attempted operative removal. At necropsy, limited to the head, a tumor 3 cm. in diameter was situated beneath the left lobe of the cerebellum and anterior to the pons. It was firmly attached to the basilar meninges and extended from the foramen magnum to the posterior clinoid processes. It eroded the base of the skull at the foramen magnum and the inner portion of the petrous ridge on the left. The spinal accessory nerve was stretched over the tumor and the left vertebral artery lay in a groove in the tumor. Microscopically, the neoplasm was poorly differentiated. The cells in a few areas showed vacuolation of the cytoplasm and a slight amount of intercellular mucin (Fig. 5).

Case 6

A 40-year-old white female complained of severe pains in the left side of the head beginning 5 months before admission. Following this, she developed a staggering gait, blurring of vision, tinnitus, numbness of the left side of the face, weakness of the left arm and leg, difficulty in swallowing, and stiffness of the tongue. Physical examination showed a staggering gait, with cerebellar signs on the left and bilateral nystagmus on lateral gaze. There was left facial paresis and slight tactile loss in the distribution of the left 5th cranial nerve. There was absence of the left supra-orbital reflex, with neuroretinitis, and concentric contraction of the visual fields. A clinical diagnosis of a large left cerebellar tumor, possibly a left acoustic tumor, was made. At operation, on elevating the left cerebellar hemisphere, a tumor was seen pushing the 7th, 8th, 9th, 10th, and 11th cranial nerves outward. The capsule of the gelatinous granular neoplasm was incised and as much of the tumor removed as possible. Erosion of the basilar portion of the occipital bone was present. The patient was hospitalized for 2 years following the operation. About 1 year before her second admission she left the hospital. Severe headaches, projectile vomiting, weight loss, and difficulty with ambulation preceded the second admission. She died immediately following a partial surgical removal of a large

recurrent chordoma. At necropsy, the neoplasm covered the anterior surfaces of the pons and medulla oblongata, extending from the partially destroyed posterior clinoid processes to the lower margin of the medulla oblongata. Destruction of bone at the base of the skull and laterally of the petrous portions of the temple bones was present. Both cavernous sinuses were invaded. All cranial nerves excepting the first and second were surrounded by neoplasm. The microscopic structure of the chordoma was similar for both surgical specimens.

Case 7

A 16-year-old white female developed severe headaches which occurred daily about 3 years before admission. Vomiting and double vision developed about 3 months before admission. Physical examination showed papilledema, cerebellar signs, absent abdominal reflexes, horizontal nystagmus to the right and left, spastic tetraparesis with weakness more pronounced on the right, disturbance of posterior column function on the right, bilateral damage of spinal nerve roots in the uppermost cervical segment, and involvement of 11th and 12th cranial nerves. A myelogram showed obstruction at the inferior margin of the atlas. A clinical diagnosis of a lesion at the upper cervical cord and lower medulla oblongata was made. At operation a pale, gelatinous, granular, tumor mass was found anterior to the cervical cord and medulla. Severe arterial hemorrhage was encountered and the patient died during the operative procedure. No necropsy was performed. The microscopic sections of the surgical specimen showed masses of vacuolated cells in a mucinous matrix.

Case 8

A 17-year-old white female complained of headache and blurred vision for 4 months prior to admission. Three weeks before admission she developed nausea and vomiting. Tinnitus in the right ear and staggering gait as well as hoarseness and vocal weakness developed about 2 weeks prior to admission. There was moderate weight loss, with general weakness and fatigue. Physical examination showed bilateral papilledema with questionable abnormal downward convergence and questionable proptosis of the left eye. Hoarseness, diminished gag reflex, and difficulty in swallowing were present. The tongue showed atrophy on the left as well as deviation to the left. A positive Romberg test and ataxic gait were present. Roentgenologic study showed a space-occupying lesion in the region of the pons displacing the third ventricle upward and the aqueduct of Sylvius backward. Visual field studies showed enlargement of the blind spot on the left and a right

homonymous hemianopsia. At operation a large tumor lay on the anterior and left lateral aspects of the pons. Decompression was carried out and the patient died the following day. Necropsy was not performed. Microscopic examination of the surgical specimen showed some cytoplasmic vacuoles but more striking nuclear vacuoles. A moderate amount of intercellular mucin was present in a few areas (Figs. 6 and 7).

Case 9*

A 52-year-old white female fell from a chair about 2 years before admission. Severe head pain and stiffness of the neck followed. Prior to death she developed difficulty in swallowing, paresthesia of the hands, and weakness of the upper and lower extremities. Physical examination in the year prior to death showed a tender, cherry-sized thickening over the second cervical vertebra. Spastic tetraparesis was present and involvement of the 11th and 12th cranial nerves. Throughout her 3-year illness the patient was thought to have cervical Pott's disease. At necropsy a neoplasm was found to extend from the medulla oblongata through the foramen magnum and to involve the first and second cervical vertebrae extensively. The neoplasm grew within the spinal dura mater. Microscopically, the large vacuolated neoplastic cells lay in pools of mucin.

Case 10

A 31-year-old male had symptoms about 8 or 10 years before the first excision. These consisted of a nasalized voice and nasal obstruction with a gradually developing mass in the throat. In 1925, an intra-oral excision was performed followed by local application of radium. For about 9 years he was symptom free. A second excision was performed in 1935 and a recurrence appeared shortly afterwards. In 1937 a nasopharyngeal cyst was removed with some relief of symptoms. The third and final excision performed in 1940 was not considered complete because invasion of the body of the atlas was thought to be present. At the time of the final excision the neoplasm consisted of an ovoid mass, 2 cm. in diameter, in the midline, elevating the posterior pharyngeal wall. It pressed the soft palate forward. The mucous membrane was not involved. Roentgenograms showed irregularity of the anterior surface of the body of the atlas. Repeated checkups for 4 years showed no evidence of recurrence and the patient died 5 years following the final excision. Death was attributed to cardiac failure and Bright's disease by the patient's home physician. Speci-

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mens from two of the excisions (1925, 1940) showed vacuolated and non-vacuolated cells in a mucinous matrix. There was no necropsy. This is the only instance in the entire series of malignant cases in which a clinical cure may have been obtained.

Case 11

A 50-year-old white male complained of attacks of pain in the back, radiating to the right arm, weakness of the right arm and leg, incontinence of urine and feces, and loss of vocal strength. These symptoms had a gradual onset over a period of 1 year before admission. Sensory loss from the upper thorax down and progressive respiratory difficulty characterized the period immediately before admission. Physical examination showed respiratory distress. He talked in a whisper. The sensory level corresponded to the cervicotrigeminal junction on the left and to the third cervical segment on the right. Papilledema, lingual paralysis, inability to raise his head, flaccid paralysis of the lower extremities, and absence of deep tendon reflexes were present. Clinical diagnosis was that of a space-occupying lesion from the first to the third cervical vertebra. The patient died 2 days following partial surgical removal. At necropsy, after removal of the vertebral laminae, a subperiosteal tumor was found extending from C 1 to C 5 on the left posterolateral wall of the vertebral bodies. From C 2 to C 4 a tumor mass narrowed the lumen of the spinal canal by about two-thirds. The prosector thought that the neoplasm arose from the left posterolateral aspect of the intervertebral disk between C 1 and C 2. Microscopically, the neoplasm was composed of vacuolated and non-vacuolated chordoma cells. Some mucin formation was present and the cells tended to be poorly differentiated, with many hyperchromatic nuclei.

Case 12

About 1 year before admission an 18-year-old female noted onset of pains in the chest and back. Gradually paralysis of the legs developed. Physical examination revealed irregular pupils, nystagmus, left facial weakness, increased deep tendon reflexes, bilateral abortive ankle clonus, absent vibratory sense in the lower extremities, and semi-flaccid spastic paralysis of the lower extremities. Roentgenograms of the dorsal and lumbosacral vertebrae were normal. A clinical diagnosis of spinal cord tumor at C 8 was made. At operation an extradural tumor 4 cm. long and 2 cm. wide was found. The seventh cervical nerve was stretched over the tumor. After removal, the posterior surface of the body of the seventh cervical vertebra was found to be roughened and the surgeon noticed that the tumor did not come from

an intervertebral disk. Extension of the tumor through the seventh intervertebral foramen was noted. Death occurred 13 months following the surgical procedure. No necropsy was performed. Before death, roentgenograms of the chest showed a soft tissue tumor mass in the left apical region. It was sharply circumscribed and extended into the thoracic inlet for about 3 cm. from the vertebral border. The trachea was displaced to the right. There was destruction of the bodies of the first and second thoracic vertebrae and the first thoracic body had slipped downward and over the second. Microscopically the tumor consisted of vacuolated chordoma cells invading dense connective tissue. There was very little mucinous matrix. Nearly every cell was vacuolated.

Case 13

A 62-year-old male noted postural low back pain which was intermittent at first but became constant, with radiation down both legs to the feet about 6 months before admission. The pain was associated with increasing constipation and pencil-sized stools. Physical examination showed a palpable descending colon. There was hypo-esthesia, more marked on the right, in the lateral sacral regions corresponding to the fourth sacral nerves. Diminished Achilles reflex and decreased vibratory sense in the lower extremities were more marked on the right. There was tenderness over the lumbo-sacral region. Roentgenograms showed gross irregularity, predominantly on the left, of the dorsal portion of the first lumbar vertebral body and pedicle. A myelogram showed complete block at the level of the first lumbar vertebra. The clinical diagnosis was a lesion at the conus terminalis. Chordoma was considered, as was neoplasm of the colon. At the first operation a tumor lying anterior to the posterior spinal ligament was described as being the size of a large olive and was thought to be coming from the intervertebral disk between the first and second lumbar vertebrae. It had produced angulation of the cord by backward pressure. Complete excision was thought to have been obtained and tumor substance in the vertebral body was cauterized. Figure 8 shows the microscopic picture. About 1 month following operation an erythema dose (650 r. to an 8 by 10 cm. field) of x-ray irradiation was given the tumor site along the spine. Three months later a similar dose was given to the same field. The patient was reasonably well for about 2½ years following these procedures. He then developed severe pain in the back and received a third x-ray treatment of 600 r. to a 20 by 15 cm. field over the original tumor area. His pain became much worse following the x-ray therapy and a second operation was performed. The recurrent chordoma invaded the lower lumbar vertebrae and extended into

the paravertebral muscles. A large amount of tumor was excised in order to decompress the cauda equina and upper conus terminalis. The second surgical specimen was similar to the first. Following this, the patient experienced little benefit and was admitted to a series of hospitals elsewhere. It is believed that he died a few months later because of the neoplasm.

Case 14

A 47-year-old white male had had his coccyx removed 11 years prior to admission for an undetermined condition. Eight years before admission he had had an injury to the spine. His present illness had its onset 3 years before admission with pain in the back, the right hip and leg. There was also a weight loss of 45 lbs. and increasing constipation. Physical examination revealed pain on thumping the lower spine, a list to the right on standing, absent knee jerks, sluggish Achilles reflexes, and diminished vibratory sense in the lower extremities. Roentgenograms showed a destructive process of the posterior and left side of the third lumbar vertebral body. A left psoas bulge was present and widening of the neural canal at this point. The clinical diagnosis was that of spinal cord tumor at the second and third lumbar vertebrae. At operation a bluish gray, gelatinous tumor was found extradurally on the left lateral aspect of the spinal cord, extending anteriorly. It invaded the body of the third lumbar vertebra. After the operation an erythema dose of deep x-ray therapy was given over the operative site. Following these procedures the patient remained bedridden because of severe pain. About 10 months later a bilateral anterolateral chordotomy was performed at the level of the seventh cervical spine. This procedure was unsuccessful and intractable pain continued. A second chordotomy was performed about 1 year later. A large tumor mass developed over the lower lumbar spine and the patient died about 1 year and 5 months following the second chordotomy. Necropsy was not performed. Microscopically, cords of chordoma cells were present lying in pools of mucin.

Case 15

A 57-year-old man gave a history of having injured his sacrum about 36 years prior to admission. About 2 years before admission he noted onset of constipation with development of ribbon stools. A mass developed over the sacrum followed by sciatic pain. Fecal incontinence and urinary frequency developed about 2 weeks prior to admission. Physical examination revealed a hard mass over the lower sacrum, severe sciatic pain on palpation, careful gait, left leg held semiflexed, absent knee jerks, sensory loss over the sacrum, and relaxation of the

anal sphincter. Ankle jerks were diminished. Saddle anesthesia with hypo-esthesia over the genitalia were present. A retropubic mass could be palpated and also a mass filling the hollow of the sacrum was found on rectal examination. Roentgenograms revealed a destructive lesion arising from the fourth and fifth sacral segments, more on the left than the right. The clinical diagnosis was that of a sacral tumor involving the third and fourth sacral nerves on the left and the fourth and fifth sacral nerves on the right. At operation, the sacrum, coccyx, posterior tumor mass and the tumor mass filling the pelvis were excised with great difficulty. Neoplasm extending upward into the spinal canal was also excised as were involved laminae. About 7 weeks following operation the patient died, probably of spinal meningitis. Necropsy was not performed. Microscopically, the neoplasm consisted of cords and strands of chordoma cells in a mucinous matrix. Some contained large vacuoles. Many of the nuclei were hyperchromatic.

Case 16

A 65-year-old white male had onset of pain in the back about 4 years before admission. The pain radiated to the right leg. Constipation and ribbon stools gradually developed. Physical examination revealed an orange-sized mass in the hollow of the sacrum which was tender to palpation. Vibratory sense was diminished at the ankles. Spinal puncture showed a complete block. Roentgen examination revealed widened space between the sigmoid colon and the sacrum. A calcareous shadow was present along the posterior surface of the lumbosacral articulation. The clinical diagnosis was chordoma or neurofibroma. At operation a 10 by 10 by 6 cm. tumor, attached to the upper anterior surface of the sacrum and occluding the sigmoid colon by pressure, was removed. It was thought at the time of operation that complete removal was not obtained, as there was infiltration in the region of the sigmoid colon. A recurrence developed and the patient died of neoplasm about 3 years later. Necropsy was not performed. Microscopically the tumor was composed of vacuolated chordoma cells with a mucinous matrix.

Case 17

A 53-year-old white female noted pain in the lower back with radiation in the sciatic areas of both thighs, which had had its onset about 1 year before admission. There were numbness and paresthesia over the buttocks and in the lower extremities. She had been confined to bed by pain for 8 months prior to admission. Bowel and bladder incontinence was present. Physical examination showed pain on pressure

over the coccyx. There was no rectal mass. Saddle anesthesia with extension to right buttock, weakness of both legs, and hyperactive knee jerks were present. Absent ankle jerks and a right Babinski sign were noted. Roentgenograms showed destruction of the sacrum below the first sacral segment with apparent expansion of the sacral body. The clinical diagnoses were those of conus terminalis lesion and chordoma. A biopsy was performed, but no treatment was instituted. Two years later roentgenograms showed destruction of the entire sacrum with additional destruction of the last lumbar vertebra and the medial portions of the ilia. Prior to death the patient developed a large mass in the sacral region. Microscopically, the biopsy showed bulky fragments of vacuolated chordoma cells.

Case 18

A 53-year-old white male had his sacrum injured by a blow 1 year prior to developing a lump on the sacrum. In 2 years the lump grew to the size of an egg. No other symptoms were recorded. Operative removal was performed but recurrence developed and in 5 years an orange-sized mass could be palpated in the hollow of the sacrum. It presented also beneath the anal skin. A second excision was performed. In addition, the patient received three x-ray treatments of 200 r. each over the sacrum and coccyx. Eleven years later a third excision was performed. The baseball-sized tumor mass over the lower sacrum involved gluteal muscles on either side. Three years later a fourth excision was carried out of a tumor measuring 6 by 8 cm., which covered the remaining portion of the sacrum and extended into the sacral foramina. The patient died of bronchopneumonia following operation. At necropsy, masses of neoplasm cells were found microscopically in the operative area. The several surgical specimens showed no essential variation and consisted of vacuolated chordoma cells in a mucinous matrix.

Case 19

A 76-year-old female injured her back 4 or 5 years prior to admission, while lifting a patient. She had onset of pain in the sacrum about 2 years before admission. This was exacerbated by straining at stool. Some constipation was present. Physical examination showed a sausage-shaped mass lying in the hollow of the sacrum, which could be felt on rectal palpation. Roentgenologic examination showed anterior displacement of the rectosigmoid junction, with non-visualization of the lower end of the sacrum and coccyx. The clinical diagnosis was that of a sacral tumor. At operation the eroded sacrum and coccyx and tumor were removed en bloc. The moderately well encapsulated tumor

mass weighed 105 gm. and measured 7 by 8 by 4.5 cm. (Fig. 11). Microscopically, it showed a variable pattern but some areas had vacuolated cells in a mucinous matrix (Figs. 9, 10, 12, and 13). Careful study of the margins of the specimen showed that the neoplasm extended to the line of excision in at least one area. The patient continued to have severe pain following operation and was bedridden most of the time. No recurrent neoplasm could be palpated on two physical examinations. She died about 1 year following the surgical excision. Necropsy was not performed.

Case 20

A 68-year-old white male was admitted because of dysuria and severe pain in the back. Physical examination showed a grape-fruit sized mass attached to the left portion of the sacrum. On rectal examination there was a large retrorectal mass involving the left side of the sacrum. Roentgenograms showed a destructive lesion involving nearly all of the sacrum and expanding its cortex. The cortex was broken through in numerous places. The clinical diagnosis was sacral chordoma. At operation the large tumor mass was determined to be inoperable and biopsy was performed. Microscopically, the neoplasm consisted of vacuolated and non-vacuolated chordoma cells with some mucinous matrix. Infiltration of bone was present. The patient was discharged and a follow-up report several months later indicated that the patient was having great pain, and could feel the mass in the sacral area.

Case 21

A 26-year-old female had a tumor removed from the sacral area, which had been diagnosed as a malignant cystadenoma. Eight years later a large encapsulated tumor in the same region was removed and on microscopic examination showed chordoma cells in a mucinous matrix, with moderate nuclear pleomorphism and hyperchromatism. Later history was not obtained.

Case 22

A 59-year-old male had noted a tumor over the sacrum and coccyx for 7 years. This became tender and produced a throbbing pain in the sacral area. He had had an operation of unknown nature on the rectum 2 years before admission. Physical examination showed a semifluctuant, orange-sized mass attached to the coccyx. It was tender and extended to the posterior margin of the anus. Neurologic findings were not noted. Roentgenologic examination showed complete destruction of the coccyx and invasion of the terminal segments of the sacrum.

Clinical diagnosis was sarcoma of the coccyx. The neoplasm was excised and the patient received a single dose of 650 r. over the oper-ative site. Four years later he had a large recurrent neoplasm measuring 12 by 12 by 5 cm., occupying the lower two-thirds of the sacrum and the space between the gluteal muscles. Neoplasm could be felt on rectal examination. Roentgenologic examination at this time showed destruction of the sacrum extending to the third sacral segment. In the year prior to death he received deep x-ray therapy consisting of 2000 r. to one sacral port and 1800 r. to two anterior ports over the inguinal regions. Following this there was relief of pain for 2 or 3 months but no change in the size of the tumor mass. Subsequently 1800 r. to two posterolateral ports cross firing the base of the tumor was given without noticeable effect. Necropsy was not performed.

DISCUSSION

The 18 cases reported here illustrate nearly all of the clinical and pathologic features associated with malignant chordomas in the several anatomical locations in which the tumors arise. Tables I and II summarize pertinent clinical and anatomical data.

TABLE I

Location, Age, Sex, and Duration of Malignant Chordoma Cases

Case no.	Age	Sex	Location	Approximate duration of illness
5	13	M	Blumenbach's clivus	2 yrs.
6	40	F	Blumenbach's clivus	4 yrs.
7	16	F	Blumenbach's clivus	3½ yrs.
8	17	F	Blumenbach's clivus	4 mos.
9	52	F	Cervical vertebrae 1 and 2	3 yrs.
10	31	M	Cervical vertebra 1 and nasopharynx	25 yrs.
11	50	M	Cervical vertebrae 1 and 2	1 yr.
12	18	F	Lower cervical and uppermost thoracic vertebrae	2 yrs.
13	62	M	Lumbar vertebrae 1 and 2	5 yrs.
14	47	M	Lumbar vertebra 3	6 yrs.
15	57	M	Sacral segments 4 and 5	2 yrs.
16	65	M	Upper sacrum	7 yrs.
17	53	F	Sacrum	7 yrs.
18	53	M	Lower end of sacrum	19 yrs.
19	76	F	Lower end of sacrum and coccyx	3 yrs.
20	68	M	Sacrum	Still living after 3½ yrs.
21	26	F	Sacrum	At least 8 yrs., no follow-up
22	59	M	Coccyx	10 yrs.

TABLE II
Therapy Used in Malignant Chordoma Cases

Case no.	Surgical excisions and explorations	X-ray and radium therapy
5	Death during suboccipital craniectomy, 1925	None
6	(a) Suboccipital craniectomy, 1930 (b) Death following suboccipital craniectomy for recurrent chordoma, 1933	None
7	Death during suboccipital craniectomy and cervical laminectomy, 1939	None
8	Death 1 day following surgical decompression	None
9	None	None(?)
10	(a) Intra-oral excision, 1925 (c) Intra-oral excision, 1935 (d) Incision, nasopharyngeal cyst, 1937 (e) Intra-oral excision, 1940; not considered complete but patient apparently cured	(b) 50 mg. of radium in nasopharynx for 12 hrs.; 2 mos. later 50 mg. of radium in nasopharynx for 14 hrs.; 1925
11	Death following cervical laminectomy, 1942	None
12	Death 3 mos. following cervical laminectomy, 1930	None
13	(a) Lumbar laminectomy, 1935 (c) Death 10 mos. following lumbar laminectomy, 1938	(b) Erythema dose over tumor site along spine, 8 x 12 cm. field, 650 r., following operation; repeated 3 mos. later (d) 20 x 15 cm. field, 600 r., single dose, 1938, following operation
14	(a) Lumbar laminectomy, 1934; death 3 yrs. later (c) Chordotomy for pain, 1935 (d) Chordotomy for pain, 1936	(b) Single erythema dose of x-ray irradiation over operative site, 1935
15	Death from meningitis 1 mo. following surgical removal of sacral chordoma, 1934	None
16	(a) Enucleation of sacral chordoma, 1939 Death 3 yrs. later	(b) X-ray therapy elsewhere, 1940(?)
17	(a) Biopsy, 1942, of sacral chordoma; death 6 yrs. later (b) Bilateral cervical chordotomy for pain	None
18	(a) Excision of sacral chordoma elsewhere, 1925 (b) Excision, 1930 (d) Excision, 1941 (e) Excision, 1944; death 1 mo. later	(c) Following operation 200 r. to 15 x 15 cm. field over sacrum, 6/9/30; 200 r. to 15 x 15 cm. field over coccyx, 6/10/30; 200 r. to 15 x 15 cm. field over coccyx, 6/11/30
19	Excision, 1949; death about 1 yr. later	None
20	Biopsy, 1950	None
21	(a) Excision of sacral chordoma about 1932 (b) Excision, 1941	Not known
22	(a) Excision of sacral chordoma, 1935; death 5 yrs. later	(b) Single dose of 650 r. over operative site, 10 x 12 cm. field, 1935 (c) 1939, three ports: two anterior, one posterior; 2000 r. to sacral port, 1800 r. to anterior ports over inguinal regions (d) 1940, 1800 r. to two posterolateral ports, cross firing the base of the tumor

Except for the cases of Koritzky[15] and Rubaschow[16] involving the maxillary and mandibular alveolar ridges, and the more plausible case of Alezais and Peyron[17] in the left occiput outside of the skull, all chordomas have been found at the base of the skull or along the vertebral column. In this series 4 arose from Blumenbach's clivus; 3 were upper cervical including the nasopharyngeal chordoma; 1 was low cervical or upper thoracic; 2 arose from the lumbar region; 7 from the sacrum; and 1 from the coccyx. The dorsal spine is not represented definitely in this group of cases but origin in the dorsal spine has been reported.[18]

Ten of the patients were males and 8 were females. The ages at onset varied from the second decade to the eighth. The disease has been seen in the fetus[19] and in young children.[20]

The duration of the illness varied from 4 months to 25 years depending primarily on the location and secondarily on the treatment. In general, tumors of the base of the skull and cervical region cause death several years sooner than chordomas involving the lumbar, sacral, and coccygeal regions. The more often the tumor can be partially excised, the longer the life of the patient. Survivals of several years without surgical treatment were seen in cases 9, 17, and 20, indicating the slow growth of the neoplasm. This is in marked contrast to the brief course of 4 months of case 8, which arose from Blumenbach's clivus.

The prognosis of malignant chordoma is hopeless with the possible exception of such rare cases as case 10, a nasopharyngeal tumor, which is considered as demonstrating a possible cure. The inability to remove small lesions completely because of location, as in the lumbar region in case 13, or the failure of the surgeon to do more than enucleate a circumscribed accessible mass, as in cases 18 and 19, reduces the probability of effecting a cure. It is doubtful if clival and cervical chordomas which involve the central nervous system can be cured by present methods. The more radical surgical approaches advocated by Mixter and Mixter[21] and Coley[22] are definitely indicated in sacrococcygeal chordomas.

The treatment, other than surgical removal, has been x-ray therapy and radium application. Limited x-ray therapy was given to several of the patients but only case 22 involving the coccyx received adequate dosage and pain decreased for a few months without change in size of the neoplasm. Radium was applied to the nasopharyngeal area in case 10 after the first excision of the tumor.

Lack of radiosensitivity has been the experience of most x-ray thera-

pists with this tumor. Recently Wood and Himadi[23] indicated some usefulness for x-ray therapy in a large series of cases.

The clinical history, physical findings, and course of the illness are all related closely to the gross pathologic aspects of the tumor. Pain was the most prominent feature of the disease in these patients and was present in most at the outset. The invasion of bone, central nervous system, and nerves accounts for this finding. Special signs and symptoms related to the region were an important part in the clinical picture with intracranial and cervical chordomas. The nasopharyngeal case presented an otolaryngologic problem.

Case 12 showed an intrathoracic mass but it was not associated with special symptoms. The lumbar and sacral chordomas presented the findings and symptomatology of lesions of the conus terminalis and cauda equina. An additional problem was seen in the sacrococcygeal cases, with obstruction of the sigmoid colon and rectum simulating intraluminal tumors of these organs. Palpation of these masses through the rectal wall was of diagnostic value. Prior to death, an externally visible tumor mass developed with several of the lumbar, sacral, and coccygeal chordomas.

Metastases have been described in lymph nodes, liver, and other organs as a late manifestation of chordomas,[24,25] but none were found in these patients upon only 5 of whom were necropsies performed.

Grossly, the chordoma is a gelatinous gray-white tumor growing in alveolar masses (Figs. 11 and 12). Hemorrhage and necrosis in the tumor are common. Confusion with mucin-forming adenocarcinoma, chondrosarcoma, and myxoma has been a diagnostic problem in gross interpretation.

Microscopic diagnosis in all of these cases depended in the final analysis upon finding tumor cells with one or more cytoplasmic vacuoles lying in a mucinous matrix. Most of the variants described by Alezais and Peyron[17,26] were illustrated in this series.

Etiologic factors and the details of the histogenesis remain obscure. Some workers have thought that trauma plays an etiologic rôle because it is often referred to in case histories and because of the experimental work of Ribbert mentioned previously.[13,26] The rôle of trauma is not convincing in the cases reported here.

Origin of the malignant chordoma from either nucleus pulposus cells of the intervertebral disk, from benign chordomas, and from notochordal rests has been considered, but detailed proof of the source of the neoplasm has not been given. The neoplasms, in cases such as those presented here, are usually too large at the time of operation or necropsy to be of much help in studying histogenesis. Case 13 is a

possible exception since the surgeon described it as an olive-sized tumor coming from an intervertebral disk in the lumbar spine. If all or some malignant chordomas arise in benign neoplasms, the problem still remains whether the benign lesions come from nucleus pulposus cells or from notochordal rests. In general, the opinion about cell rests in recent years has been against the idea that they have some special propensity for undergoing neoplastic transformation in contrast to normally located cells.[27]

Case 4 of the benign chordoma series is of great value since the coccyx was sectioned serially and the neoplasm was seen to come out of the intersegmental disk. The tissue of the disk that would correspond to a nucleus pulposus showed rests of nucleus cells which were partly calcified. These 2 cases lend slight support to the idea that some chordomas originate from cells of a nucleus pulposus.

Resolution of the problem of histogenesis may be possible by the experimental approach.[14]

CONCLUSIONS

Four cases of benign and 18 of malignant chordoma have been reported. One of the benign chordomas was discovered in a surgical specimen of the coccyx.

No symptoms or clinical findings were attributable to the benign chordomas.

One of the 18 patients with malignant chordoma is still alive with neoplasm. One had no follow-up. The remaining 16 are all dead; one nasopharyngeal case may have been cured by repeated excisions and radium applications.

In 2 cases, one malignant and one benign, there was suggestive evidence that the neoplasm arose from nucleus pulposus cells of the intervertebral disk and from an intercoccygeal disk.

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DESCRIPTION OF PLATES

PLATE 120

FIG. 1. Case 1. Mass of physaliferous cells lying next to the circular sinus of the pituitary gland. Hematoxylin and eosin stain. × 250.

FIG. 2. Case 4. Partially calcified nests of physaliferous cells in the intercoccygeal disk. Hematoxylin and eosin stain. × 200.
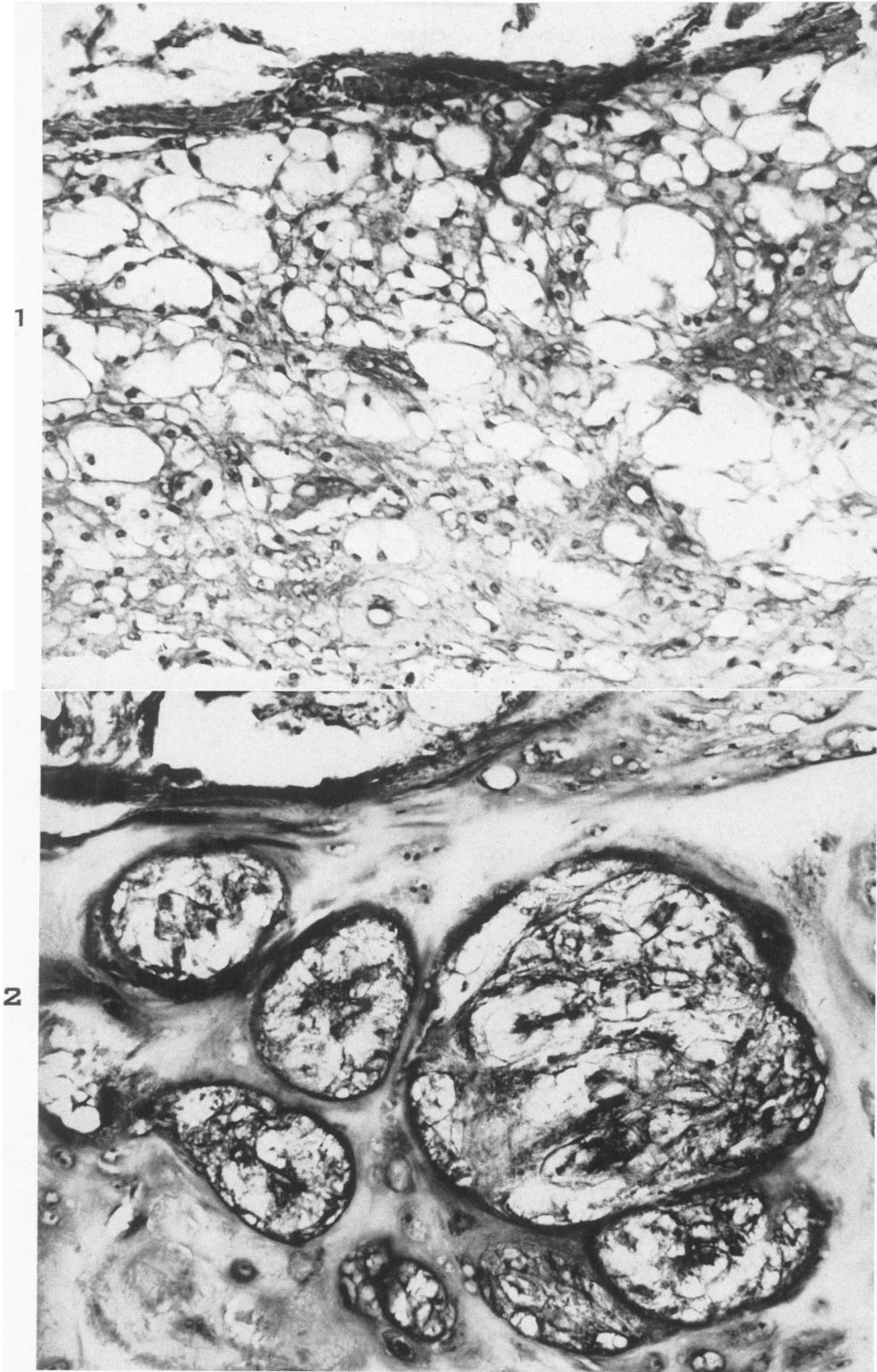
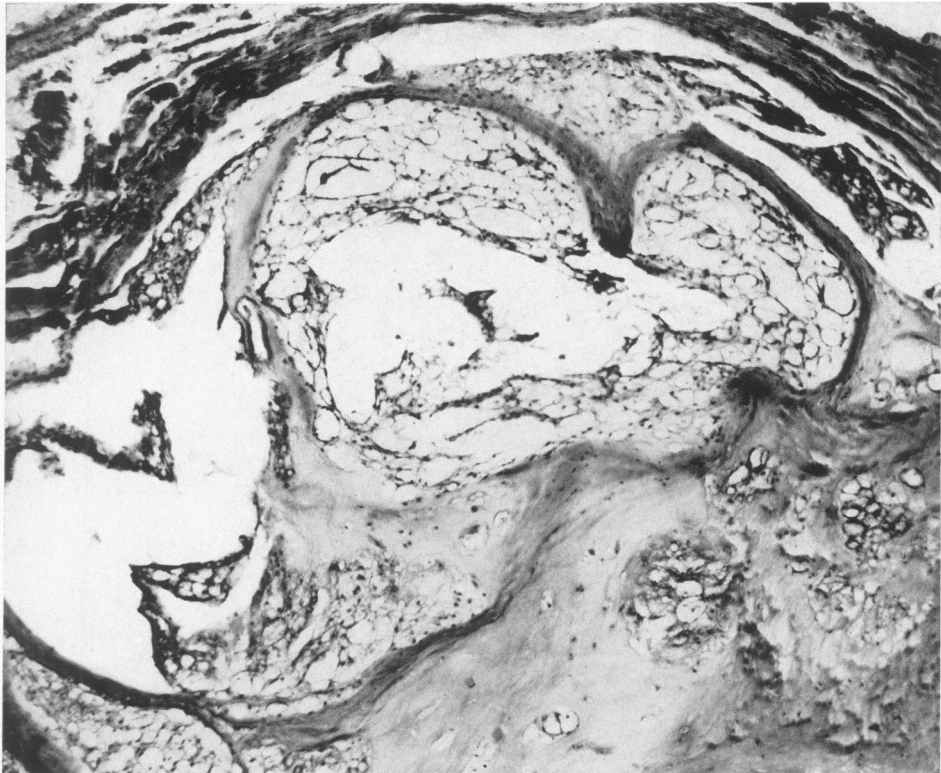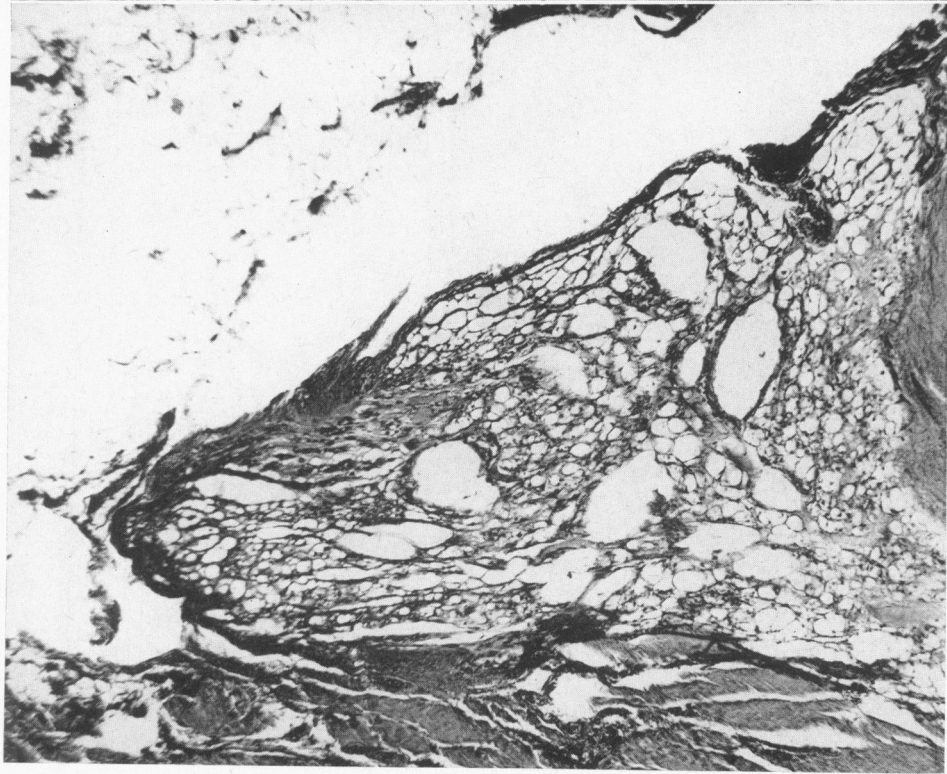Congdon   Benign and Malignant Chordomas

PLATE 121

FIG. 3. Case 4. Large nests of physaliferous cells on the surface of the first inter-coccygeal disk. Hematoxylin and eosin stain. $\times$ 75.

FIG. 4. Case 4. Main tumor mass growing in dense connective tissue adjacent to the coccyx. Hematoxylin and eosin stain. $\times$ 65.
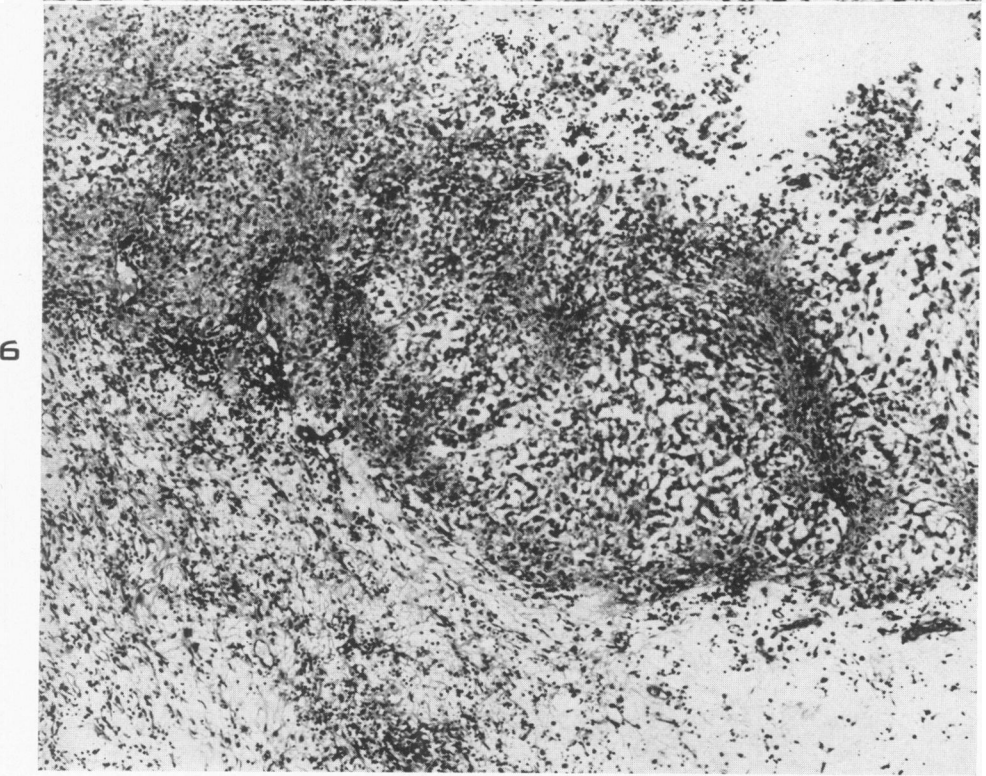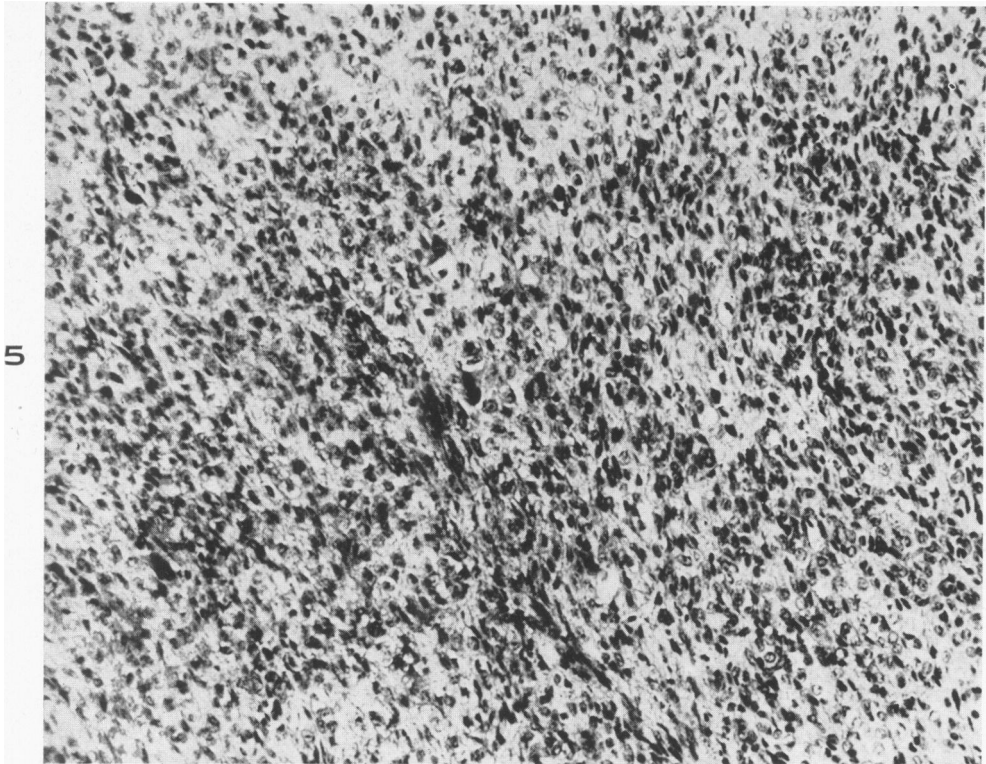
Congdon   Benign and Malignant Chordomas

PLATE 122

FIG. 5. Case 5. Poorly differentiated chordoma of the clivus. The sarcomatous appearance is evident. Hematoxylin and eosin stain. × 250.

FIG. 6. Case 8. Mass of chordoma cells. Cords of cells are separated by a mucinous matrix. Hematoxylin and eosin stain. × 80.

Congdon

Benign and Malignant Chordomas

PLATE 123

FIG. 7. Higher power view from case 8 showing cells with vacuolated cytoplasm. Solitary vacuoles are present in the nuclei of many cells. Hematoxylin and eosin stain. $\times$ 200.

FIG. 8. Case 13. Cords of neoplasm cells in a mucinous matrix. Some show cytoplasmic vacuoles. Hematoxylin and eosin stain. $\times$ 250.
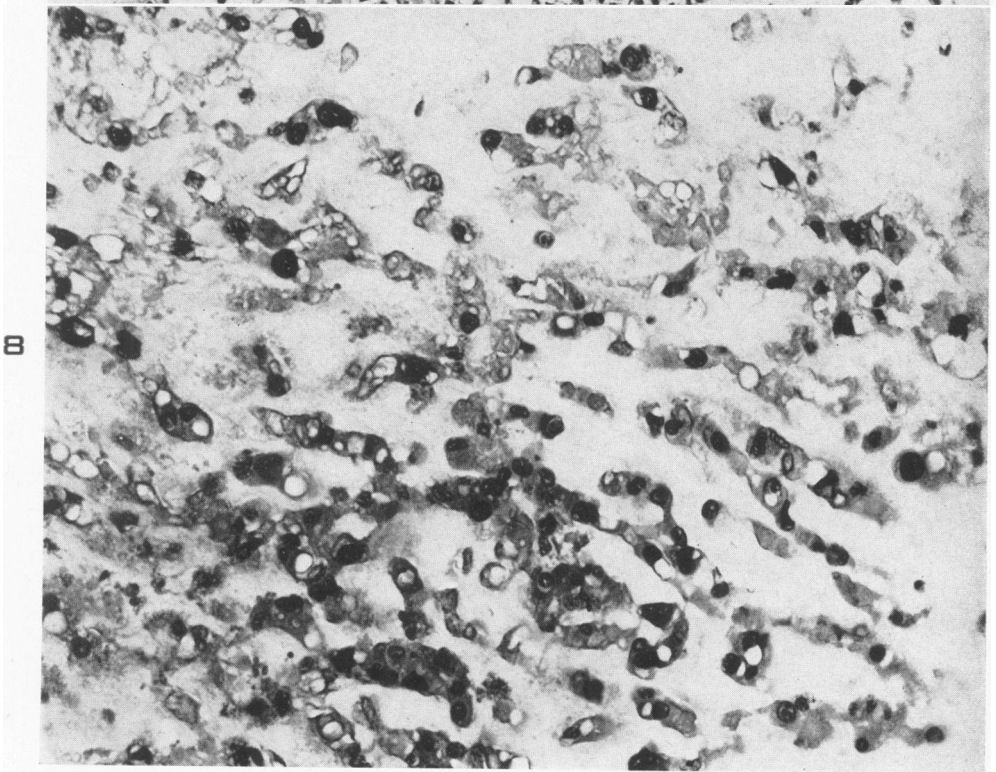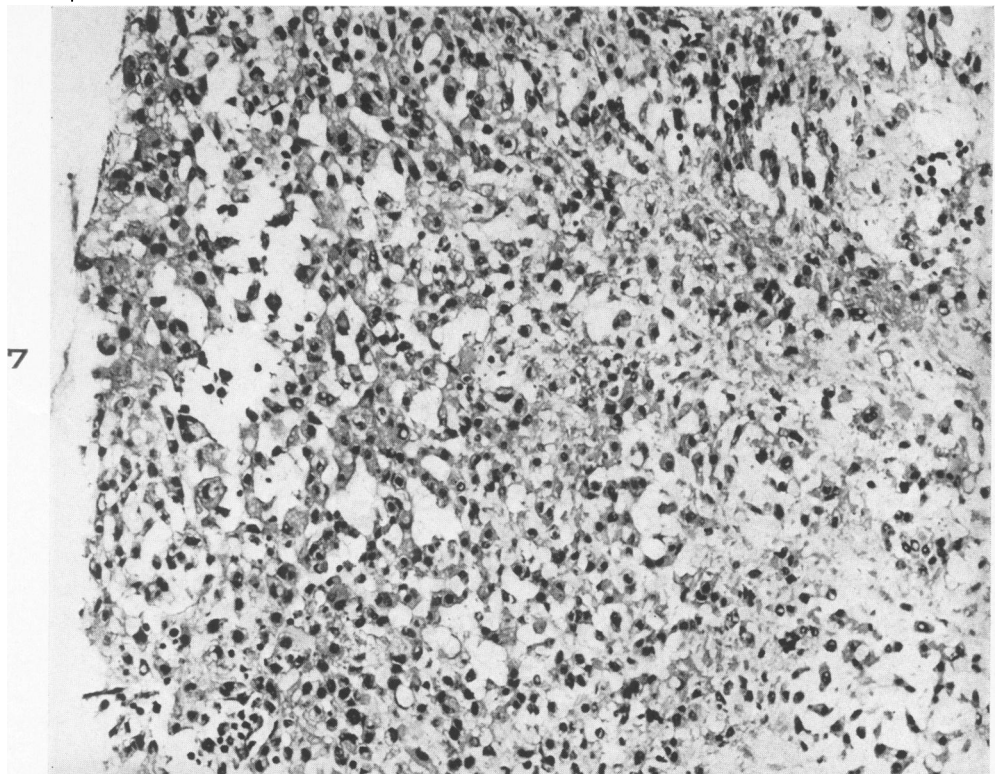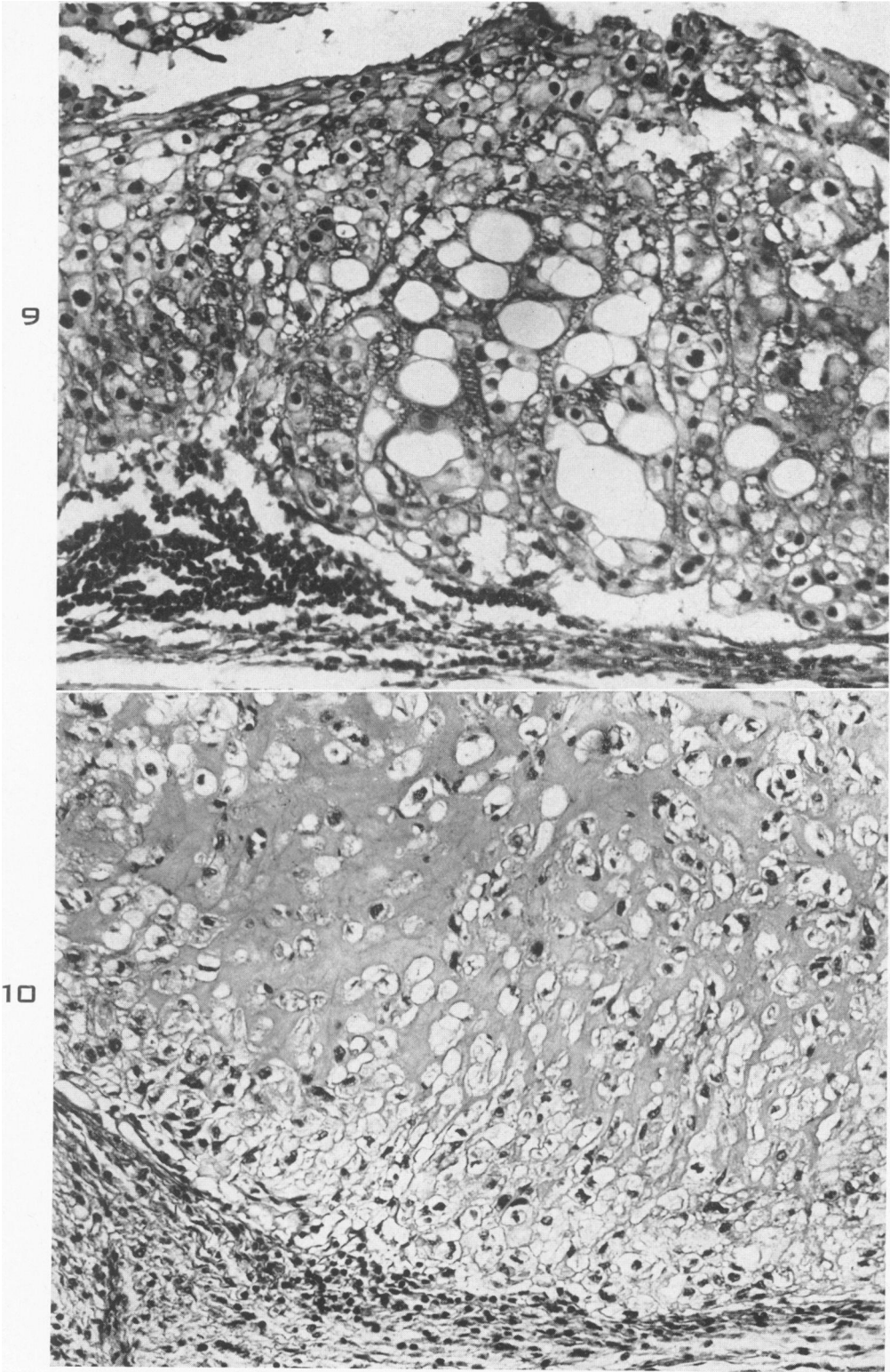
Congdon   Benign and Malignant Chordomas

FIG. 9. Case 19. Area of vacuolated cells from the edge of the main tumor mass. Hematoxylin and eosin stain. × 250.

FIG. 10. Another field from the same section as Figure 9, in which the mucinous matrix is dense, giving the tumor a cartilage-like appearance. Hematoxylin and eosin stain. × 200.
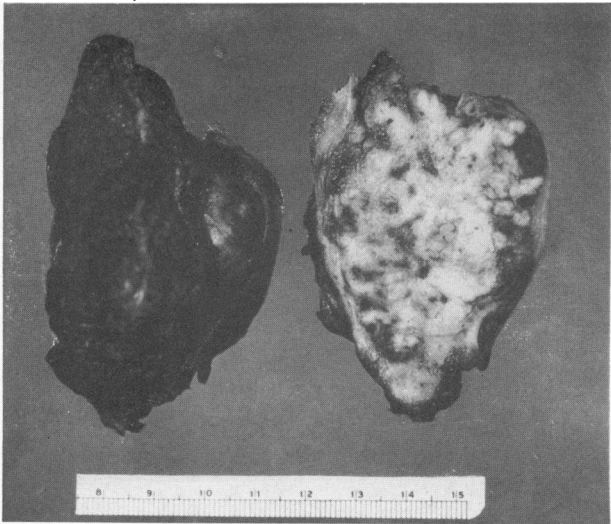
9

10

Congdon   Benign and Malignant Chordomas
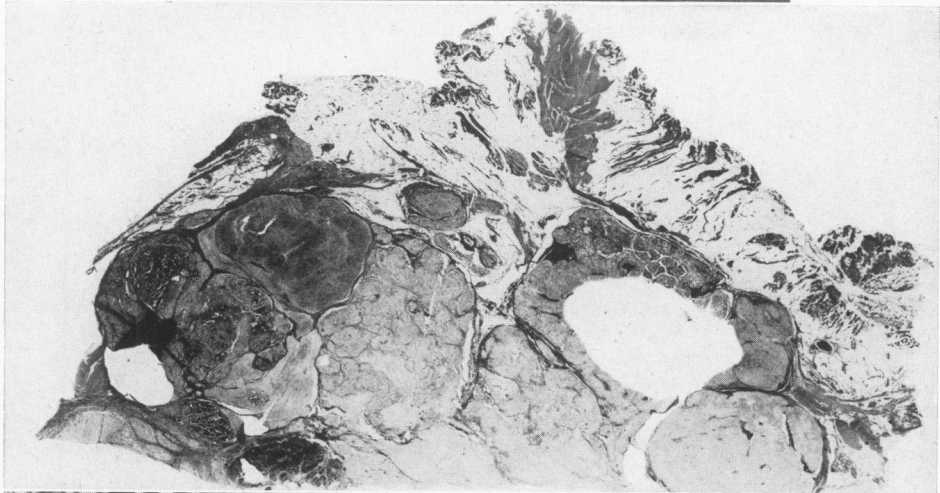
FIG. 11. Cut surface of tumor removed surgically in case 19. There is encapsulation in some portions of the circumference and none in others.

FIG. 12. Low-power view of the section from which Figures 9, 10, and 13 were taken. Of note are the alveolar growth pattern of the tumor and lack of encapsulation. Hematoxylin and eosin stain. × 10.

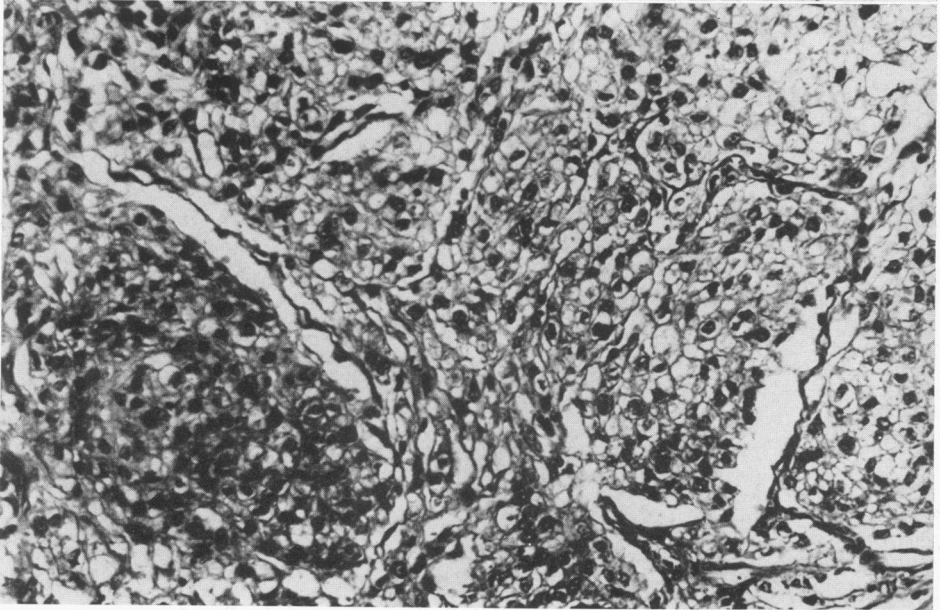FIG. 13. A third area from the same section as Figures 9 and 10 showing a fibrillar portion of the neoplasm. Hematoxylin and eosin stain. × 170.